

Partial Lipodystrophy and Successful Pregnancy Outcome

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Partial lipodystrophy is a rare disorder, characterised by lipoatrophy and insulin resistant diabetes¹. Other features include hyperlipidemia, acanthosis nigrans, hepatomegaly and mesangiocapillary glomerulonephritis. Successful outcome of pregnancy is uncommon. We report a case of favourable outcome in a female with partial lipodystrophy.

Case report

A 31 year old female presented to the ante-natal clinic at 9 weeks of her pregnancy. She was diagnosed as having diabetes mellitus, 5 years previously and was on oral hypoglycemic agents. She also had hyperlipidemia on previous laboratory investigations. She conceived soon after her marriage. There was no family history of diabetes or other abnormalities. When seen initially, she had almost complete loss of subcutaneous fat over her face, upper limbs and trunk. She had well developed musculature and mild facial hirsutism. She had 3 hyperpigmented areas on her trunk. There were no xanthomas or xanthelasma. Her BP was 120/80 and other cardiovascular examination was normal. There was no hepatomegaly. Initial ultrasound showed a single fetus at 9 weeks gestation. Her initial investigations showed Hb 13.4 gm/dl, HDL cholesterol 0.64 mmol/l, cholesterol 7.61 mmol/l and triglycerides 7.10 mmol/l, fasting blood glucose 11.9 mmol/dl random blood glucose 15.7 mmol/dl. Her renal and liver functions, serum testosterone and urinalysis were normal. She was started on insulin twice daily. Her repeat ultrasound examination at 16 and 29 weeks was normal. Her total weight gain during pregnancy was 8 kg. During pregnancy her insulin requirement increased gradually till she was receiving 96 units daily in two divided doses. She was monitored closely and her pregnancy was uneventful till 38th week when she suddenly developed preeclampsia with BP 190/110 mmHg, pedal oedema and proteinuria. Ultrasound of fetus showed discrepancy between biparietal diameter and fetal length consistent with 32 weeks and 36 weeks respectively. She was admitted and blood pressure was controlled with intravenous hydralazine, following which she underwent caesarian section and delivered a normal baby of 3.5 kg weight. Immediately following delivery her insulin requirement dropped to 16 units in 24 hours and her blood pressure was controlled without treatment.

Discussion

Partial lipodystrophy is characterized by lipoatrophy and insulin resistant diabetes. Apart from ketosis resistant diabetes and total loss of body adipose tissue, other features include hyperlipidemia, hepatomegaly, acanthosis nigrans, mesangiocapillary glomerulonephritis and increased basal metabolic rate with normal thyroid function². Partial lipodystrophy is more common with loss of adipose tissue in parts of the body and associated with some or all the above features. Certain types of partial lipodystrophy are inherited as autosomal dominant traits^{3,4}. The loss of adipose tissue in these forms usually spares the face. The most common type of partial lipodystrophy is progressive or cephalothoracic lipodystrophy resulting in a progressive loss of subcutaneous fat in the upper half of the body including face. This disorder is not inherited and affects women more than men⁵. Pregnancies in women with partial lipodystrophy carry a high risk of intrauterine death^{6,7}. Intrauterine growth retardation is also seen in these cases and the exact cause not known but most occur in the third

trimester of pregnancy. Two case reports of successful outcome of pregnancy have also been reported⁸. The patient we described had features consistent with partial lipodystrophy loss of subcutaneous fat over her face and upper trunk, in association with diabetes mellitus and hyperlipidemia. Her history indicated that the onset of this disease was in early adult life. She had a non-eventful pregnancy until 38 weeks, when she developed significant pre-eclampsia with ultrasound evidence of growth retardation. Her outcome as described was successful. As most patients described in the literature develop problems late in third trimester as did our patient, we feel early elective delivery should be advocated for these patients. Partial lipodystrophy may be associated with successful pregnancy outcome but these patients require very close monitoring.

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